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ARCHIVES OF PEDIATRICS

A MONTHLY DEVOTED TO THE

DISEASES OF INFANTS AND CHILDREN

JOHN FITCH LANDON, M.D., Editor

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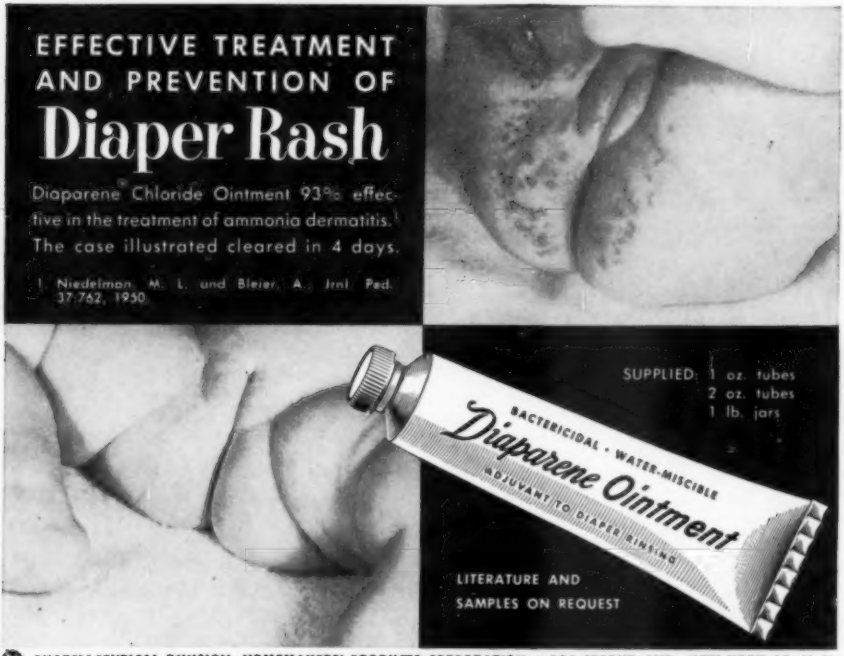
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1. Grayzel, H. G., Heimer, C. B., and Grayzel, R. W.: New York St. J. Med. 53:2233, 1953. **2.** Heimer, C. B., Grayzel, H. G., and Kramer, B.: Archives of Pediatrics 68:382, 1951. **3.** Behrman, H. T., Combes, F. C., Bobroff, A., and Leviticus, R.: Ind. Med. & Surgery 18:512, 1949. **4.** Turell, R.: New York St. J. Med. 50:2282, 1950. **5.** Marks, M. M.: Missouri Med. 52:187, 1955.

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DIAGNOSIS OF THE MILD FORM OF CHRONIC RHEUMATIC HEART DISEASE IN CHILDREN AND YOUNG ADULTS*

HERMAN ROBBINS, M.D., F.A.C.C.
Ridgewood, N. J.

The diagnosis of chronic rheumatic heart disease can and should be made clinically. This applies to the mild as well as to the severe forms of the disease. While it is comparatively simple to recognize the latter, the milder forms presenting no outstanding familiar physical signs or symptoms, remain largely unrecognized. The criteria for diagnosis, to be presented here, are based on the observation of hundreds of cardiac children during their acute stage on the ward and then followed for years in the Cardiac Clinics.

Rheumatic involvement of the aortic valve can be readily diagnosed. The characteristic diastolic murmur in its usual location, the nature of the pulse, the unusually low diastolic blood pressure and the heart contour showing left heart enlargement, make the diagnosis not too difficult. However, in the great majority of rheumatic hearts, the only valve involved is the mitral valve. Among these may be found the most severe as well as the mildest forms of the disease. One of the important things to be remembered in considering the diagnosis of chronic rheumatic heart disease is the fact that rheumatic manifestations of the acute stage

* Based on observations by the author while Chief of Pediatric Cardiology, 1938-1955, at the Flower and Fifth Avenue Hospitals and the Metropolitan Hospital, New York, N. Y.

may have been so very mild as to escape notice, or, may have been noticed and misunderstood. In suspected chronic cases, a careful questioning may elicit a positive history. However desirable a definite history may be, a diagnosis may be made without it.

First of the diagnostic signs for consideration is the apical systolic murmur. In chronic rheumatic mitral disease, this murmur may be loud and transmitted or it may be faint and localized in the region of the cardiac apex. If faint, it may be brought out on exercise, or the systolic murmur may be absent altogether. There are systolic murmurs near the apex that are non-organic or due to a congenital cardiac defect. It is thus evident that an apical systolic murmur, regardless of its intensity, cannot itself serve as a means of diagnosis. The only murmur that can be considered characteristic of mitral disease, is the presystolic murmur. The failure to recognize the mild form of chronic rheumatic mitral disease can be traced to the failure to recognize the early presystolic murmur. It is still unfortunately being taught that a presystolic murmur can be identified by its rumbling nature. It so happens that there is no rumble characterizing the presystolic murmur of childhood. Instead, it is identified by what seems to be an accentuated, prolonged or hesitant first heart sound. This includes the murmur and the first sound. To best hear this murmur, the patient should be placed in the recumbent position. If he has had rheumatic fever, the presystolic murmur of the chronic phase, should be sought for and invariably will be found localized just to the right of the cardiac apex. In addition to this characteristic murmur, further auscultation will reveal the accentuation of the second heart sound in all other locations, wherever the heart sounds are heard. The examination should be made before exercising the patient. Examination after exercise, will produce an accentuation of the first sound in all precordial areas. Thus, in mild cases having an early presystolic murmur, this is apt to disguise an important diagnostic sign.

The blowing diastolic murmur heard at the apex, occurring together with a loud systolic is indicative of comparatively severe damage to the mitral valve. It is easily recognized and does not fall into the category of the mild type of case under discussion here.

When a presystolic murmur has been found, the diagnosis of mitral disease may be confirmed by viewing an anteroposterior

chest x-ray. It must be emphasized that the criteria for determining cardiac enlargement in adults cannot as a rule be used in the case of children. When this difference is not appreciated, only the serious cases with much enlargement will be recognized. Among normal children, heart size shows considerable variations. This contributes to the difficulty of recognizing slight and moderate over-all enlargement. Rheumatic involvement of a cardiac valve, throwing as it does an extra amount of work on certain individual chambers, causes these chambers to enlarge. This type of enlargement produces an abnormal cardiac contour, and it is the recognition of the abnormality of the contour that supplies the confirmation of the diagnosis by auscultation. In chronic mitral disease the x-ray picture in the anteroposterior view shows a straightening of the left cardiac border. The transverse diameter need not show enlargement.

As already stated, a history of acute rheumatic fever is not a requirement for a diagnosis of a chronic rheumatic heart. However, a history though inconclusive will direct the physician to a thorough search for the presence of the early presystolic murmur. In the mild form of chronic rheumatic carditis, neither the cardiogram nor the oblique chest x-ray offer any additional conclusive evidence for diagnosis.

This paper has presented a simple clinical procedure and criteria for the recognition of the mild form of chronic rheumatic heart disease in children and young adults. The acute rheumatic infection, in late years, has taken on a milder course with consequent less residual cardiac damage. One cannot overemphasize the need for recognition of the early and mild form of this disease, in children at school, in young adults preparing to begin their life work, in youths at the armed services induction centers. Acute recurrences of rheumatic fever are possible, not only in childhood but in adult life as well. The need for prophylactic care is quite evident.

STUDIES ON NORMAL SERUM ALKALINE PHOSPHATASE IN EGYPTIAN INFANTS AND CHILDREN*

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and
EISA, E. A., M.SC., PH.D.
CAIRO, EGYPT.

*From the Pediatric and Chemistry Departments, Ein Shams University, Cairo, Egypt.

It has long been known that serum alkaline phosphatase is higher in children than in adults. However, the studies in this field are remarkably few and the findings published by different authors and from different localities show wide variations. We intended, therefore, to study the normal levels of serum alkaline phosphatase in Egyptian newborns, infants and children. This is of prime importance as a basis for studying their variations in disease, especially as one notices that in recent years the literature on the significance and interpretation of phosphatase levels in various bony diseases, endocrine disturbances and hepatic affections, is rather rapidly increasing.

EXPERIMENTAL WORK

The study was performed during the summer of 1956. It was carried out on 106 cases, of which 30 cases were newborns, 24 were infants between one month and two years and 52 were children between two and fourteen years. The cases studied were chosen from among healthy newborns in the obstetric section, and from healthy newborns, infants and children, attending the outpatient of Abbasia Children's Hospital with their brothers or sisters suffering from some illness. The height and weight for each case were recorded, then 5 cc. of blood were taken for the estimation. All precautions were taken to prevent blood haemolysis.

The method of estimation adopted is a combination of Bodansky's Method[1, 2] and that of Fiske and SubbaRaw[3]. The principle of the procedure depends on the incubation of serum with buffered glycerophosphate at pH 8.6 for exactly one hour, after which the serum proteins are precipitated with trichloroacetic acid. The protein-free filtrate, containing the liberated phosphate is treated with an acid molybdate solution to form phosphomolybdic acid, which is reduced by the addition of one, two, four aminonaphthol sulfonic acid reagent to produce a blue colour, the intensity of which is proportional to the amount of phosphate present.

The Bodansky unit for phosphatase activity is the difference between the inorganic phosphate content of the incubated and control samples under conditions mentioned above, and expressed in mg. of phosphorus per 100 ml. of serum.

THE RESULTS

The results of the present work are illustrated in the following tables and curves.

TABLE I. It shows the age, sex, height and weight of the cases studied, with their normal alkaline phosphatase levels.

Av. Age	No. of Cases M.	F.	Av. Height in Cms.	Av. Weight in kilos.	Av. Serum Alkaline Phosphatase in B.U.
Dys.					
1	2	–	49	3.22	4.5
2	–	2	49	3.00	4.4
3	1	1	49.5	3.75	4.4
4	2	4	49.5	3.60	4.3
5	1	2	51.0	4.3	3.7
7	2	2	50.0	3.5	4.5
8–28	6	5	51.5	4.05	4.5
Mos.					
1–3	4	2	53.0	4.3	4.25
4–6	1	2	59.0	6.4	5.8
7	2	1	65.0	5.5	5.9
8	1	1	64.5	7.75	5.8
10	2	–	66.5	8.55	5.95
12	1	1	69.0	8.42	6.15
15	2	–	76.0	11.25	7.35
18	1	1	78.0	11.25	8.25
24	1	1	81.0	11.60	8.45
Yrs. Mos.					
2–3	5	4	93.0	14.4	7.24
4–5 8	8	6	104.0	18.3	5.2
6–7 6	1	6	114.0	23.4	5.01
8–8 3	2	3	121.0	25.0	4.90
9–9 3	2	2	125.0	28.0	4.65
Yrs. Mos.					
10–10 3	1	2	130.0	30.1	4.3
11–11 5	2	3	136.0	31.5	5.24
12–12 6	1	2	141.0	36.2	5.60
13–14	–	2	146.0	40.0	5.70
	51	55			

TABLE II. The average values of normal serum alkaline phosphatase in the first week, first 2 years and in the years from 2 to 14.

Average Age		Phosphatase Units (Bodansky)
1	day	4.5
2	days	4.4
3	"	4.4
4	"	4.3
5	"	3.7
7	"	4.5
1	month	4.5
2	months	4.2
4	"	5.3
5	"	5.8
7	"	6.0
8	"	5.8
10	"	6.0
12	"	6.3
15	"	7.3
18	"	8.2
24	"	8.5
3	years	7.3
4	"	6.3
5	"	5.7
6	"	5.3
7	"	5.1
8	"	4.9
9	"	4.6
10	"	4.4
11	"	5.0
12	"	5.5
13	"	5.6
14	"	5.7

TABLE III. It shows the minimum, maximum and average values of serum alkaline phosphatase in the age periods studied.

Age Period	No. of Cases	Min. B. U.	Max. B. U.	Average B. U.
First month	30	2.5	6.0	4.0
1 mo. to 2 yrs.	24	3.5	8.7	6.0
Two yrs. to 14 yrs.	52	3.2	8.0	5.5

TABLE IV. The changes in the average levels of serum alkaline phosphatase from one period to another.

Age Period	No. of Cases	Change in Average Level	Starts by B.U.	Ends by B.U.	Average B. U.
First wk.	19	A temporary drop	4.5	4.5	4.3
First 2 yrs.	24	progressive rise	4.5	8.5	6.2
2 yrs. to 10 yrs.	39	gradual drop	8.5	4.4	5.7
10 yrs. to 14 yrs.	13	A second but less marked rise	4.4	5.7	5.2

DISCUSSION

I. From the foregoing tables and curves the following points can be noted:

1. In the ages studied, serum alkaline phosphatase levels showed no variation with regard to sex. (Table I).

2. At a given age, the normal variations of height and weight do not affect the levels.

3. The intensity of the physiological jaundice has no relation to the levels.

II. The levels in different age periods:

a. *The first week* shows the lowest levels. This result is in agreement with the very few reports done in this field, namely those of Stearns and Warweg[4], Abul Fadl and Zahira[5] and Abul Fadl and Awatif[6]. We also noticed a temporary drop during the first week. This was not recorded by other workers. The explanation for the low serum alkaline phosphatase levels during

the first week, in spite of the fact that the phosphatase content of the bone is highest just before birth may be that during that time the newborn suffers from birth shock which, no doubt, greatly diminishes the body activities, including the osteoblastic one. It can be noticed also that the temporary drop observed during the first week coincides, more or less, with the normal drop of weight, and the diminution of the body activities during that period. (Table II).

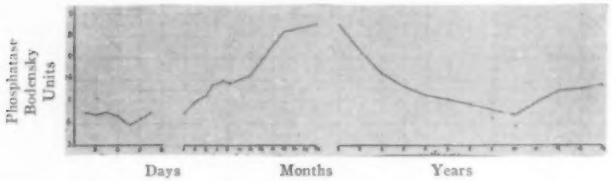

Fig. 1. A composite curve showing the consecutive changes that occur in serum alkaline phosphatase units in the first wk., first two months and from 2 to 14 yrs. of age.

b. *During the first two years,* the figures obtained show a progressive increase in alkaline phosphatase levels. This is in fair agreement with the figures found by Stearns and Warweg[4] who concluded that the activity increases after birth and remains

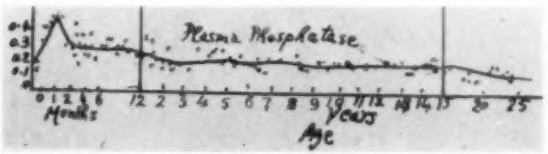

Fig. 2. A curve of a similar work done by Stearns and Warweg on American Children from Iowa City.

comparatively high, well into the second year. This is explained by the rapid rate of growth which occurs at that time. The figures of Abul Fadl and Zahira[5] for this period cannot be used for comparison, as they did not mention a sufficient number of cases to allow for a safe conclusion, and moreover, they themselves expressed some doubt regarding the normality of the cases they studied.

TABLE V. A comparative study of the normal range of variation of serum alkaline phosphatase activity during the age period from 6 months to 2½ years.

Authors	No. of Cases	Min.	Max.	Average
Klasmer (1944)	320	5.5 B.U.	15 B.U.	9.4 B.U.
Awwaad & Eisa (1957)	20	4.9 B.U.	8.7 B.U.	6.68B.U.
Abul Fadl & Zahira (1952)	16	2.4 A.U. = ±0.88 B.U.	15.5 A.U. = ±6.2 B.U.	7.00A.U. = ±2.8 B.U.

TABLE VI. A comparative study of the normal range of variation of serum alkaline phosphatase activity in children of 2 years to 10 years of age.

Authors	No. of Cases	Min.	Max.	Average
Bodansky & Jaffée (1934)	19	3.1 B.U.	10.7 B.U.	7.3 B.U.
Awwaad & Eisa (1957)	41	3.2 B.U.	8.7 B.U.	5.5 B.U.
Abul Fadl & Zahira (1952)	25	3.0 A.U. = ±1.2 B.U.	15.9 A.U. = ±6.3 B.U.	6.0 A.U. = ±2.4 B.U.

TABLE VII. A comparative study of the normal range of variation of serum alkaline phosphatase activity in children over 10 years of age.

Authors	No. of Cases	Age Period	Min.	Max.	Average
Bodansky & Jaffée (1934)	6	10-15 yrs.	5.3 B.U.	13.1 B.U.	9.0 B.U.
Awwaad & Eisa (1957)	11	10-14 yrs.	3.9 B.U.	5.7 B.U.	5.3 B.U.
Jules et al. (1947)	50	10-12 yrs.	6.6Sh.U. = ±2.2 B.U.	17.1Sh.U. = ±5.7 B.U.	11.1Sh.U. = ±3.7 B.U.

The period from six months to two and a half years was further considered with regard to its importance as a common age incidence for rickets. The present findings show that the figures for the Egyptian infants are a little bit lower than those of Klasmer[7] for infants in Jerusalem as shown in Table V. Although the findings of Abul Fadl and Zahira[5] are in fair agreement with Klasmer's figures, they are still very low, since the Armstrong units they used are two and a half times greater than Bodansky units (Table V.)

c. The period from two to ten years of age shows a declining variation, the average values starting from a maximum of 8.5 B.U. and ending with a maximum of 4.4 B.U. This decline is in agreement with the findings of Stearns and Warweg[4] (Fig. 2) and those of Bodansky and Jaffée[8] (Table VI). The previous Egyptian findings of Abul Fadl and Zahira[5] show a similar decline but still with comparatively very low figures, since their maximum average was 10.8 A.U., which nearly corresponds to about 4.0 B.U. A summary for the above mentioned statements is given in Table VI.

d. From ten years to fourteen years of age, we noticed a slight rise, the average levels starting from 4.4 and ending in 5.7 B.U. (Table IV & Fig. 2). This second rise compares well with the findings of Bodansky and Jaffée[8] (1934) and Jules et al[9] (1947) as shown in Table VII.

The fact that Abul Fadl and Zahira[5] did not reach a similar conclusion regarding the alkaline phosphatase content of blood serum of normal Egyptians during this period is most probably due to the small number of cases they studied. The rise in this period is easily explained by the great activity and maturation which occurs then.

SUMMARY AND CONCLUSIONS

1. 106 cases of normal Egyptian infants and children were studied regarding the alkaline phosphatase content of their blood serum during that age period.

2. The effect of sex, age, height and weight on the phosphatase content was observed.

3. From the average levels found at different age periods, a composite curve was made. This curve illustrates the consecutive changes in serum alkaline phosphatase levels from one period to the next. It is a double rise curve; the first rise is a marked one and occurs in the first two years of life. The second rise is less marked and occurs between ten years and fourteen years. It is noted that the rises parallel the activity and rate of growth during the same age.

4. The results obtained in the present work were discussed and compared with those found by previous research in that field.

5. Generally speaking, the figures for Egyptians in the ages studied are slightly lower than those mentioned for similar ages elsewhere. This may be due to variations in climatic, dietetic and racial conditions.

REFERENCES

1. Bodansky, A.: J. Biol. Chem., 99, 197, 1932.
2. Bodansky, A.: J. Biol. Chem., 101, 93, 1933.
3. Fiske and SubbaRaw: J. Biol. Chem., 66, 375, 1925.
4. Stearns, G. and Warweg, E.: J. Biol. Chem., 102, 749, 1933.
5. Abul Fadl, M. A. M. and Zahira, H. A.: Proc. Pharm. Soc. Egypt., 34, 89, 1952.
6. Abul Fadl, M. A. M. and Awatif, R.: Gaz. Egypt. Pediat. Ass., 2: 292, 1954.
7. Klasmer, R.: Am. J. Dis. Child., 67: 348, 1944.
8. Bodansky, A. and Jaffée H. L.: Arch. Int. Med., 54: 88, 1934.
9. Jules, et al.; J. Lab. & Clin. Med., 32: 194, 1949.

DEATH FROM CAT SCRATCH DISEASE. (Monatsschr. Kinderh., 102:232-237, April 1954). Gräff reviews the present status of knowledge on cat scratch disease, pointing out that it is a lymphadenitis, which, depending on the site of invasion of the causal factor, presumably a virus, may become manifest as a cutaneoglandular, a tonsilloglandular, or an oculoglandular type. According to Mollaret, the diagnosis can be verified with the aid of a complement fixation test some weeks after the appearance of the glandular process. Although it has been generally accepted that the disease runs a favorable course, terminating in spontaneous cure, Gräff reports the case of a 30-month-old boy who died after an attack of cat scratch disease.—*J.A.M.A.*

STAPHYLOCOCCUS ALBUS AND SYSTEMIC SENSITIZATION*

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The prolonged febrility which may mark the process of joint sensitization is often devoid of demonstrable hematogenous bacterial agents. This means that one will often deal with a puzzling systemic disorder which is typified by its resistance to antihistaminic, antibiotic and adrenal cortical substances. The disorder, then, self-limited in its quality, will eventually clarify itself and show its reaction in cynovial membranes at which time the effect of adrenal cortical agents is often dramatic.

The role of certain strains of streptococci in the pathogenicity of arthritis is not questioned. The necessity for differentiating the signs and symptoms of rheumatic fever from those of infectional arthritis seems to be open to serious doubt. The writer has observed four cases of severe residual joint deformity in children with typical rheumatic heart disease.

A remarkable instance of rheumatic fever due to hemolytic staphylococcus albus was observed by the author. The illness began with acute tonsillitis and generalized erythema. Four days later, septicity was marked. Staphylococcus albus was recovered from the throat and blood stream. Two weeks later, a mitral murmur was noted which later changed to the complex of murmurs indicative of a double mitral lesion. There was multiple joint involvement in a migratory pattern. This patient showed no residual involvement of the joints after a three month period.

CASE REPORTS

C.S. was 13 months old at the onset of her illness. Fever was abrupt in its onset on July 2, 1956. There was a generalized papular urticaria which was intensely pruritic. Physical findings, other than the presence of vasomotor rhinitis, were nil. The skin disorder later assumed an angry erythematous appearance aggravated by a myriad of scratches.

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This infant weighed seven and a half pounds at birth and was markedly hypertonic. She did not respond to the use of milk substitutes but improved rapidly when thick cereal feedings with homogenized milk were given.

The Hb was 9 grams per hundred cubic centimeters of blood, the RBC were 2.8 million and WBC 24,000 per cubic milliliter of blood. The urine was negative. Nose and throat cultures were reported positive for Staphylococcus albus, hemolytic, coagulase positive. The sedimentation rate was 20 mm. per hour (normal level 11 mm.). A minimal patchy right basal pneumonia was present. The spinal fluid was totally normal. Treatment was begun with Ilotycin* in dosage of 200 mg. every 6 hours with penicillin 600,000 units intramuscularly (IM.) every 12 hours. Meticorten was given in dosage of 5 mg. every 6 hours. Cortone cream was used locally. Staphylococcus albus, hemolytic, coagulase positive was recovered from blood cultures and found to be sensitive to both penicillin and Ilotycin. Antistreptolysin titre was 125 Todd units.

Six days after treatment was begun, the blood cultures became negative, but the septicity of the child was not altered. ACTH was used in dosage of 5 mg. every 6 hours IM. and elixir of pyribenzamine was given in dosage of a half dram every 6 hours orally. Two blood transfusions were given. Relief from the pruritis and lessening of the urticaria was present for transitory periods during depression of fever.

Desquamation became apparent during the second week of illness and was most marked over the feet and hands. Bilateral myringotomy became necessary but no specific organism was recovered from the serosanguinous fluid released. Febrility was present each night to the level of 104 degrees. The degree of splenic enlargement was moderate but the degree of adenopathy was extreme, particularly in the left axillary region where one gland could be felt, filling the entire axillary fossa. Chloromycetin was given for six days without effect.

A change in diet, including use of goat's milk, did not bring about improvement in the urticaria which became intense with each spike of fever. Periodic blood transfusions were necessary because of anemia. Abdominal distension became a problem and was controlled by enemas and the insertion of a colon tube.

* Supplied by the Eli Lilly Co., Indianapolis, Indiana.

One month after release from the hospital, September 1956, the temperature curve finally dropped to normal and remained so though no treatment was being given. The splenomegaly and adenopathy receded completely. The patient suddenly developed rapid swelling of the wrists, ankles, interphalangeal and intertarsal joints. Pain on motion of these joints was marked.

The joint swelling had disappeared by December 1956, and her weight had increased to twenty-six and one-half pounds. The blood count was normal at the time of examination. She developed a small furuncle on the arm during January 1957 which was followed by scattered urticaria in the area. The infection cleared rapidly without treatment.

In retrospect, the child can be regarded as having suffered from dermatitis exfoliativa due to Staphylococcus albus with the urticaria signifying the onset of sensitization and disappearing with recession of the fever which marked the end of the active sensitizing process.

Fig. 1. C. S. Showing swollen fingers and wrists.

T.C., a 14 year old girl, was admitted to the St. Francis Hospital on January 25, 1954. She had developed a mild upper respiratory infection which was followed in 48 hours by pain in the left lower chest and which was aggravated by cough and deep inspiration. The cough became progressively worse, and, at the time of admission her temperature was 101.6 degrees, orally.

The child was tall and asthenic in build with extensive acne vulgaris involving the face, neck and shoulders. The blood pressure was 110/70 mm. of Hg. Dullness and decreased breath sounds were found over both lung bases. The heart was not en-

larged and no murmur was found. Marked hepatic tenderness was noted and the liver was enlarged. A routine chest roentgenogram was interpreted as indicative of bilateral basal pleurisy without parenchymatous involvement.

Fig. 2. Chest roentgenogram.

The hemoglobin was 10 gm. per hundred cubic centimeters of blood, the RBC 4.84 million and WBC 19,950 per cubic milliliter of blood with differential showing 88 per cent polymorphonuclear cells. The antistreptolysin titre was 200 Todd units.

A pericardial effusion was found on the fourth day of hospitalization; the anemia became more pronounced with drop of Hb. to 60 per cent of normal and RBC count of 3.37 million per cubic milliliter of blood. The sedimentation rate was 15 mm. in one hour (normal 0-9 mm.).

Septicity was marked with rise of temperature level to 104 degrees daily. Staphylococcus albus (coagulase positive) was recovered from blood cultures. The urine showed 20-30 WBC per high power field.

The patient was treated with 500 mg. of Achromycin intravenously, then 250 mg. orally every 6 hours. Meticorten was given in dosage of 10 mg. every 6 hours orally. The blood culture became negative after a period of seven days.

Respiratory oppression led to the need for an oxygen tent and the patient complained constantly of pain in the upper abdomen which caused restriction in breathing.

Response to the measures outlined was lacking, and a five day trial of streptomycin, and of bacitracin, and finally polymyxin

were carried out without effect upon the septicity. The patient began to evidence mental depression and on February 21, 1954, she was dismissed in spite of febrility. All therapy was discontinued. Six weeks later, the patient appeared to be clinically well though markedly under her normal weight. She was seen at home on April 16, 1954 at which time extensive joint swelling was found, particularly in the fingers, wrists and knees. The response to meticorten, 10 mg. every 6 hours orally, was dramatic.

T.C. was found to be sixty-two inches tall and her weight was one hundred and two pounds on September 2, 1954. The acne vulgaris was very severe. No cardio-pulmonary abnormality was found and the blood pressure was 105/65 mm. of Hg. The chest x-ray was negative. Her weight was one hundred and eighteen pounds on January 15, 1955. She was observed again on March 3, 1955 because of frank delusions and ideas of reference. Institutionalization became necessary due to mixed (catatonic and paranoid) schizophrenia.

DISCUSSION

It is often stated that there is little justification for the use of the term Still's disease to indicate the juvenile form of infectional arthritis.[1] This attitude is based upon the parallel juvenile-adult effects of severe systemic reactions and the failure of the characteristics of manifestations to modify the prognosis. The interference with process of growth in the child and the juvenile's more striking septicity[1] are not regarded as sufficient criteria to warrant employment of the term Still's disease. The same situation prevails in consideration of Felty's syndrome which is merely regarded as a variant of rheumatoid arthritis. The common denominator of the above designated diseases would be the identical pathological changes which include fibrinoid swelling and fibrinoid exudate of the synovial membrane.

The differentiation between rheumatoid arthritis and rheumatic fever is often quite impossible. Changes of rheumatic carditis in rheumatoid arthritics at necropsy have been noted by Baggenstoss and Rosenberg[2]. The high frequency of this involvement again directs attention to the fact that the two so-long widely separated entities may actually represent variations of an identical process.

The most outstanding factor involved in "Still's disease" as we have observed it has been the prolonged septicity which is essential to sensitization of the synovial membrane. In one instance, that period was six years[3].

SUMMARY

Two patients are discussed who were involved with a septic process due to Staphylococcus albus, coagulase positive. Though the blood cultures became negative, the septicity persisted and was resistant to various antibiotics and cortisone. In one instance, the arthritis which developed later responded dramatically to cortisone. The process of sensitization persisted for ten weeks in each case. The youngest patient manifested her illness by dermatitis exfoliativa due to Staphylococcus albus and generalized urticaria was an annoying symptom. The older patient showed an acute pericardial and pleural effusion as the initial symptom.

REFERENCES

1. Johnson, N. J. and Dodd, K.: Med. Cl. of N. Am., p. 459, March 1955.
2. Baggenstoss, A. H. and Rosenberg, E. F.: A. J. Path., 35:503, 1943.
3. Stadler, H. E.: ARCH. PEDIAT., 73:216-219, June 1956.

ATRESIA OF THE ESOPHAGUS WITH TRACHEOESOPHAGEAL FISTULA. (J. Florida M.A., 40:621-628, March 1954). The authors advise early diagnosis of esophageal atresia, since many infants with this anomaly can be cured by surgery. The prognosis is worsened by the baby's having been offered feedings, particularly if there is a tracheoesophageal fistula. In the four cases described by the authors, two patients survived and two died, one because of delay in diagnosis and treatment (the baby had well-developed bronchopneumonia) and the other because he was a premature baby with a poor chance for survival of surgery and because no lower segment of the esophagus could be found, thus making an anastomosis impossible. He was the only one of the four who did not have a fistula. The symptoms of this condition are excessive drooling of frothy mucus; choking, coughing, and difficult breathing; and immediate regurgitation of feedings associated with choking, coughing, and cyanosis. In diagnosis, an esophageal catheter should be passed to demonstrate obstruction, iodized oil should be injected to show atresia or fistula of the upper segment, and a roentgenogram should be made of the abdomen, air in the stomach being an indication of a tracheoesophageal fistula. Surgical management of the condition is described in detail.—*J.A.M.A.*

MYOTONIA DYSTROPHICA

Report of Four Cases with Post-Mortem Findings in Three

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and

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MYOTONIA DYSTROPHICA

This report is concerned with four siblings, two brothers and two sisters, of whom the oldest is still alive at the age of 50. Three siblings are dead. The post-mortem findings in these three cases form the basis of this study. Although there are many excellent detailed descriptions in the literature of the clinical picture, biopsies and post-mortem examinations of muscle fibres, complete autopsy reports are rather few[1-4] and it was thought that three additional reports might be worthy of presentation.

The family history of these patients is meager. There is vague information that their maternal grandmother had a chronic wasting disease. The father born in 1879 was of Italian descent, the mother was born in England of English parents. The father was an alcoholic, bad-tempered, usually out of work, and the burden of supporting the family rested on the mother's shoulders. She was said to be of average intelligence. She died of cancer of the stomach at the age of 52. Of their five children the oldest, a son, was a delinquent boy with a court record. Nothing is known of him, except that he appeared to be free from myotonic dystrophy.

Case #1. Pearl C. This patient, the second child and the first daughter, was born in 1907, when the mother was 23 years old. She is the only survivor of the four sick siblings. She is said to have had pulmonary disease in childhood. Her father attempted to rape her when she was 16 and for this reason she was taken from home and put in St. Joseph's Correctional School for Girls, but she was so troublesome that the Sisters refused to keep her and she was admitted to Wrentham State School, March 14th, 1924, at the age of 17. At the time of admission she was 143.5 cms. tall, weighed 70 kilos. She had several stigmata: her palate

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was high and narrow, her lips thick, her ears coarse with attached lobules. She was a mouth-breather, had a slight curvature of the spine and a pronounced cyanosis of the extremities. Head measurements were: circumference, 48 cms., length of the head, 16.25 cms., width, 13 cms. Her speech was lispy, her skin rough, her tongue coated. Vision and hearing were good. Pupils were round and equal, reacted to light and accommodation. Patellar reflexes were sluggish, eye grounds normal, Romberg negative, gait normal, muscular strength good, Wassermann negative, Von Pirquet test positive, pulse 72. Mentally she was retarded, her I.Q. was 61 per cent. She was quarrelsome, sly, disobedient and aggressive, careless about her appearance, profane and forward with boys. Her work was poor in quality and she worked under protest. Myotonic reaction of hands was developing very slowly and perhaps her known dislike of work was the cause of her being considered lazy, when she began to have muscular difficulty.

In 1927 she had jaundice. In 1946, the x-ray examination of the skull showed thickening of the bones of calvarium and a moderate degree of hyperostosis frontalis. The shape of the skull appeared to be normal. The pineal body was not calcified. The sella turcica was not enlarged but there was a rather unusual prominence of the clinoid processes. The petrous ridges were somewhat irregular and there was poor aeration of mastoid cells. In 1949, at the age of 42, she was found to have fine punctate lenticular opacities, especially in the left eye, on examination with slit lamp. She was diagnosed as a case of Myotonia Dystrophica.

Laboratory findings at that time were: total serum cholesterol 147 mg. per 100 cc.; serum cholesterol ester was 100 mg. per cc., or 68 per cent of total. Twenty-four hour urinary output was 1140 cc.; 24 hour urinary total creatinine was 0.30 mg. (normal value 1-2 mg.); 24 hour urinary creatine reported as creatine was 0.07 mg. (normal value 3-7 mg.).

The urine examination for albumin and sugar was negative. Repeated blood counts showed gradual development of anemia with anisocytosis, presence of macrocytes, slight stippling and normal platelets. The RBC was 3.5000.000; Hg. 13 gm.; WBC 5.700 with polynuclears 59 per cent, lymphocytes 34 per cent. In 1951, the x-ray of the chest showed congenital alterations of the upper rib structures with a short cervical rib on the left side and a long one on the right. Serum Wassermann was negative; tuberculine

test, positive; histoplasmine test, negative. The last physical examination, in 1956, showed pulse 88, blood pressure 140/100, small highly arched feet, left small toe, a hammertoe. Mild myotonic reaction in hands was present, knee jerks were absent, while abdominal reflexes were present and lively. Hands were very small with atrophied muscles. She complained of fatigue and frequent stomach upsets. Gastrointestinal (G.I.) tract series were negative. She remains in poor health.

Case #2. Vincent C. This patient, a brother of Pearl and the third sibling, was born in 1910, when the mother was 26 years old. Very little is known of his early development, except that he had influenza at the age of 8. He was a dull boy; placed in a special class at school, he made no progress and was admitted to Wrentham State School, October 9, 1923 at the age of 14.

Physical examination on admission showed a well nourished boy, 146 cms. tall, weighing 40.5 kilos, with facial asymmetry, outstanding ears, and a fractured nose. Head measurements were: circumference 51 cms., length 17.7 cms., width 14.4 cms. The tongue protruded in the midline. Pupils were equal, reacted to light and accommodation, eye grounds were negative. Vision, hearing, and sensation were not impaired. Speech was not defective. Muscular strength was good. Serum Wassermann was negative.

Laboratory findings: RBC 5.740.000, Hg. 13 gms. WBC 11.500, with polynuclears 50.5 per cent, lymphocytes 48.8 per cent. Blood platelets and red blood cells were negative.

Clinical course: The patient was undisciplined, unreliable, stealing, swearing, aggressive and homosexually inclined. His school work was very poor. Gradually, he began to lose the use of his fingers. He was very slow in everything he did, first giving the impression of being lazy, sleepy and uninterested. At the age of 18, he was reported by the matron as being so lazy "he finds it hard work to even lift his cup to his mouth when he wants a drink." At the age of 19, he had diminished deep tendon reflexes and normal superficial reflexes. Station and gait were normal, Romberg negative. Coordination tests were well performed, his muscular strength was good, and his speech was not defective; vision, hearing, and sensation were not impaired. In 1945, at the age of 35 he had a well pronounced alopecia of temporal regions, dystrophy of the arms and legs, difficulty in walking long distances, protuberant abdomen, atrophied genitals,

characteristic muscular irritability (Thomsen's sign) and a tendency to obesity.

The laboratory findings were: Urinary androgens, as androsterons, 4.8 mg. in 24 hours. Total serum cholesterol was 190 mgs. per 100 cc., and serum cholesterol ester was 131 mgs. or 69 per cent of total. Twenty-four hour urinary total creatinine, 0.80 gm. per cent; 24 hour urinary creatine as creatinine, 0.06 gm. per cent; 24 hour creatine as creatine, 0.07 gm. per cent. Several x-rays of the chest were negative. In 1949, at the age of 40, his I.Q. was 50 per cent. Slit lamp showed punctate lenticular opacities near the surface and in deeper layers. In 1950, his obesity and wasting of the muscles of hands and legs were very pronounced. In 1951, heart disturbance was noticed and he was treated for cardiac decompensation. He died July 25, 1952 at the age of 43 years.

Pathologic, anatomic findings were: Acute, diffuse hemorrhagic pancreatitis; acute diffuse gastro-entero-colitis. Marked enlargement of the entire pancreas, with diffuse fresh hemorrhagic infiltration of the pancreas, also numerous small foci of fat tissue necrosis of the pancreas. There was intense diffuse inflammatory swelling and reddishness of the entire intestinal mucosa from the duodenum to the anus, numerous partially flat confluent and extensive fresh mucosal hemorrhages of the ileo-caecal region of the appendix and of the ascending colon; numerous fresh hemorrhagic erosions and fresh minute bleedings of the stomach mucosa. Intense catarrhal tracheo-bronchitis; edema of the lungs. Hypertrophy of the heart. Sclerosis of both coronary arteries, with marked narrowness of the entire aorta. Striking hyperplasia of the thymus, and extensive hyperplasia of the thyroid gland, with diffuse colloid infiltration of the thyroid (diffuse colloidal struma). Hypoplasia of the adrenals; lipoid paucity of the adrenal cortex, and marked diffuse thickening of the adrenal capsule. Striking hypoplasia and diffuse brown pigmentation of the testes, and hypoplasia of the prostate. Hyperemia of the kidneys; dilatation and catarrhal inflammation of the kidney pelvic bilaterally; marked dilatation of the ureters. Intense dilatation of the bladder; catarrhal inflammation of the bladder mucosa. Enlargement and hyperemia of the liver. Enlargement of the spleen, and softening of the spleen pulp. Numerous concretion-like cholesterin stones of the gall-bladder. Intense diffuse hyperostosis of the calvarium.

Striking narrowness of the basal skull fossae; narrowness of the foramen magnum. Intense hyperemia of the pia, with few fresh flat pial bleedings. Marked obesity. Old adhesions between upper and middle lobes of the right lung. Moderate anthracosis of the peribronchial lymph nodes. Epicrisis.

Cause of death was the acute pancreatitis which probably developed simultaneously with gastro-entero-colitis. Hypertrophy of the heart combined with marked coronary sclerosis may have accelerated the fatal issue. The following findings are thought to be connected with the conditions responsible for the muscle disease: Hyperostosis of the skull bones, hyperplasia of the thymus, diffuse colloidal struma, adrenal hypoplasia and hypoplasia of the testicles.

Miscroscopic examination showed a variety of changes in muscle fibers. Chains of central nuclei with prominent nucleoli, atrophy of muscle fibrils with replacement by connective tissue and clumps of darkly stained basophilic nuclei. Liver showed severe fatty degeneration. Pituitary showed an increase in basophiles and wide septa through which ran numerous wide blood vessels.

Case #3. Tony C., brother of Pearl and Vincent, the fourth sibling, was born in 1911 when the mother was 27 years old. No history is available of mother's health during pregnancy or the boy's early childhood. He came to the attention of the school authorities because of his lack of progress in school. In 1923 he was admitted to Wrentham State School at the age of 12. On admission, the physical examination showed a 134 cms. tall boy, pale, rather undernourished, with protruding ears and mild kyphosis. Head measurements were: circumference 50 cms., length 17.2 cms., width 13.3 cms. Pulse 68, blood pressure 100/66. Motor functions: tremor not demonstrated, station steady, Romberg negative, gait normal, coordination good, speech not defective, tongue protruded in the midline, pupils round and equal, and reacted to light and accommodation. Superficial reflexes normal. Deep tendon reflexes diminished. Eye grounds negative. Wasserman negative. Von Pirquet test positive. I.Q. 48 per cent.

Laboratory data: RBC 6.420.000. Hg. 22.6 per cent. WBC 12.000 with polynuclears 60 per cent, lymphocytes 30 per cent and monocytes 10 per cent. RBC showed slight stippling. Platelets were normal. The eye examination with slit lamp was negative. Wassermann reaction was negative. September 1945, 24-hour androgens, as androsterons, was 6.6 mg. In 1946, serum total cholesterol

was 156 mgs. per 100 cc.; serum cholesterol ester was 116 mgs. or 74 per cent of total. Twenty-four hour urine volume was 910 cc.; 24 hour urinary creatinine, 0.66 gm.; 24 hour urinary creatine as creatinine was 0.07 gm.; 24 hour creatine as creatine was 0.08 gm.; the albumin and sugar were negative.

Clinical course: He was reported to be lazy, sleepy and apathetic for several years but in 1947 it was evident that he was losing the use of his hands, and in 1950 that he was losing the strength of his arms. His obesity became noticeable in 1950. In 1953 he was treated for cardiac decompensation, and pulmonary tuberculosis. Ptosis of left eyelid was noticed about the same time. In 1955 urinary sugar was found to be three plus and alkaline phosphate 8.2 Bodansky units. Blood fasting sugar was 221 mgs. per 100 cc., NPN. was 29 mgs. per 100 cc. He was treated with insulin and placed on a diabetic diet.

He resembled his brother Vincent in many ways, had abnormal sex habits, but was quieter in his behavior. In August 1956 he became acutely ill, had a marked cyanosis, difficult respiration and marked dyspnea. He died December 1956 at the age of 46.

Pathologic, anatomic diagnosis was: Pulmonary embolism, hemorrhagic infarct of the middle lobe of the right lung; hypertrophy of the heart; chronic bronchitis and bronchiolotis; chronic cholecystitis and cholelithiasis, degeneration of the striate muscle. Almost complete occlusion of the pulmonary artery at the level of bifurcation with large thrombotic embolic masses (straddling embolus); small conical hemorrhagic infarct in the diaphragmatic margin of the right middle lobe; moderate atelectasis of the lower lobes of both lungs; marked thickening of the bronchial mucosa with the fibrinous infiltration, especially in the left lower lobe. Rather sticky, thick mucoid exudate in the bronchi, especially in the left lower lobe. Hypertrophy of both cardiac ventricles, especially the left; marked dilatation of the right atrium and ventricle; hypertrophy of papillary muscles in both ventricles, especially the left; marked thickening of and hardening of the mitral leaflets with markedly thickened chordae tendineae. Moderate thickening of the tricuspid leaflets; numerous atheromatous plaques in the entire aorta and both coronary arteries; few lentil-sized arteriosclerotic plaques in the parietal portion of the tricuspid valve; moderate thickening of the endocardium, especially at the sub-aortic region of the left ventricle. Marked dilatation

of aortic sinuses, especially the left; moderate fenestration of the aortic valves. Marked hyperplasia of the thyroid gland (50 gms.) with diffuse colloid infiltration (diffuse colloidal struma). Marked swelling and anthracotic infiltration of the paratracheal, bifurcation, hilus, intra-pulmonary lymph nodes. Marked swelling and hyperemia of the parenchyma. Moderate diffuse induration of the liver. Two cms. long ovae-shaped chrystalline cholosterol stone in the gall bladder; marked contraction of gall bladder with marked fibrous thickening of the wall. Moderate thickening of the splenic capsule; enlargement of spleen, softening of the splenic pulp. Moderate swelling and partially scattered, partially confluent hemorrhages of the intestinal mucosa, especially the small intestine; moderate narrowing of the ileum. Normal appendix. Marked swelling and hyperemia of both kidneys; indistinct cortex medullary markings in both kidneys; marked cortical enlargement of both kidneys. Marked hypoplasia of both adrenal glands; lipoid paucity of the cortex of adrenal glands; numerous pinhead size, yellowish orange colored cortical adenomas in both adrenal glands. Dilatation of the urinary bladder. Extreme hypoplasia of both testicles (both are 10 gms.); brown pigmentation of the parenchyma of both testes; hypoplasia of seminal vesicles. Marked diffuse hyperostosis of the calvarium. Marked narrowness of the basal portion of the skull with narrowness of foramen magnum; the sinuses, bones and dura are within normal limits. Marked sclerosis of the cerebral arteries; moderate sclerosis of the brain substance; marked hyperemia of the pia and brain substance. Marked obesity. The biceps and brachialis muscles showed marked softening with whitish-pink discoloration. The deltoid, brachio-radialis, rectus abdominis, pectoralis, obliquus externus abdominalis psoas major, iliacus, quadratus lumborum, muscles of the right thigh showed no gross or microscopic abnormality, except the softening and whitish discoloration of the vastus medialis. There was marked softening, whitish-pink discoloration of the right gastrocnemium and right soleus muscles (fishy appearance).

The immediate cause of death was pulmonary embolism. Hypertrophy of the heart and coronary sclerosis could have contributed. Hyperostosis of the skull, hypoplasia of testicles, seminal vesicles and adrenal glands; diffuse colloidal struma probably connected with chronic brain condition which produced the muscular disease.

Grossly, the grain showed no changes. The gyri were well developed. The sulci are neither deep nor shallow. Pia-arachnoid was thin and transparent. The pial vessels appeared injected. The pacchionian bodies were well pronounced. The vessels at the base were thin and supple. There was a slight thickening of basal pia. Septum pellucidum was duplicated forming a well pronounced cavity. Corpus callosum was negative, grossly. Section through the optic chiasma showed a somewhat dilated third ventricle. The section through the mammillary bodies showed no gross changes in mammillary bodies, basal ganglia, Ammon's horns. In this section the third ventricle showed a more pronounced dilatation. Cranial nerve showed nothing except that the right oculomotor nerve seemed to be slightly thinner than the left. Section in front of the optic chiasma, through the middle of the frontal lobe showed no gross changes in the structure of the cortex of the white matter. Section through the temporal tips exposed the frontal horns of the lateral ventricles which appeared somewhat dilated. Occipital horns of the lateral ventricles, especially the left, were dilated. Fourth ventricle showed no changes. Medulla and cerebellum showed no gross changes.

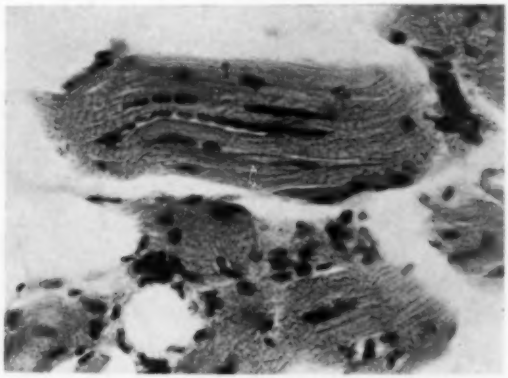

Fig. 1. To illustrate the change consisting of a production of central nuclei in parallel rows. Reduced from x800. Hematoxylin and eosin stain.

Microscopic examination: Sections of muscle tissue taken from upper arms, forearms, thighs and legs showed a great variety of changes from normal muscle fibers to complete atrophy. The central nuclear proliferation with long chain or two, three or

more parallel chains was prominent. Hyalinization, homonization and splitting of the fibers, very small fibers next to large ones

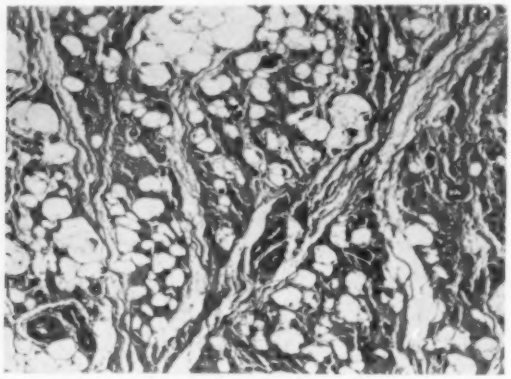

Fig. 2. Muscular atrophy. Fat and strands of connective tissue. Reduced from x800. Hematoxylin and eosin stain.

were present. There were fields where atrophic fibers were represented by clumps of dark nuclei, and bands of connective tissue and fat cells were separating atrophic muscle fibers. The "Ring-

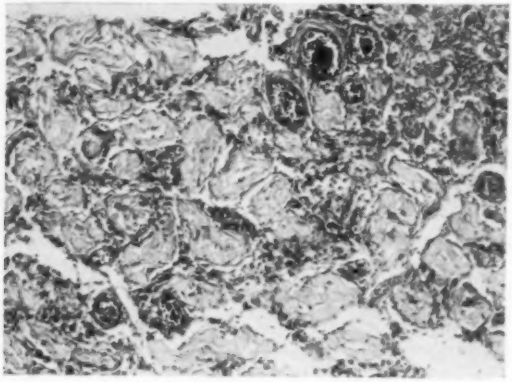

Fig. 3. Atrophy of seminiferous tubules. VanGieson stain. Reduced from x400.

binde" described by Heidenhain[12], Slauck[3], Wohlfart[4] were seen in one case only (Tony). These consisted of striated muscle fiber ring around the cross section of muscle. The liver showed very

severe fatty degeneration; testicles showed complete atrophy of seminiferous tubules with complete disappearance of spermatogenic cells and accumulation of Leydig cells. Thyroid had distended follicles filled with pink colored substance. Pituitary gland had increased number of basophilic cells forming groups of rather shapeless cells along the border of the anterior lobe. There were numerous cysts distended with colloid-like content in the pars intermedia. Lungs showed nothing of note. Myocardium showed no significant changes.

Case #4. Mary C., the youngest of the four siblings was born April 24, 1913, when the mother was 29 years old. No history was available about her early development. She was admitted to Wrentham State School, August 28, 1923, at the age of 10 years. Physical examination on admission showed a well developed and well nourished girl who was 129 cms. tall and weighed 22 kilos. She had a slight facial asymmetry, asymmetrical ears, the right much smaller than the left, high and narrow palate, mottling appearance of the skin of the extremities. Head measurements: circumference 49.5 cms., length 16.5 cms., and width 12.7 cms. Patellar reflexes were markedly diminished. Tongue protruded to the right of the median line, no tremors were observed. Pupils were round and equal, reacted to light and accommodation. Hearing and vision were apparently normal. Station steady. Romberg negative. Muscular strength was fair. Speech was not defective. Eye grounds were normal. Sensation was normal. Psychological examination: I.Q. 54 per cent. Laboratory data: RBC 4.420.000. Hg. 80 per cent. WBC 9.900. Polynuclears were 70 per cent, lymphocytes 25 per cent and monocytes 5 per cent. May 22, 1936, the x-ray of the chest showed bilateral pulmonary tuberculosis. November, 1936 x-ray showed a large cavtiy of the left apex. Acid-fast bacilli were present in sputum. The patient was very backward in her studies, used profane and vulgar language. In 1932 she had an attack of acute rheumatism. Her work was very poor and the matron considered her very lazy. She did not do anything if she could help it and her influence over other girls in the building was bad. She was transferred to the tuberculosis building. The last physical examination records high arched feet, thin lower legs, and slightly atrophic hand muscles.

She died November, 1943 at the age of 30. Cause of death: myotonia dystrophica, fibrous tuberculosis of the right lung, and

heart failure due to chronic myocarditis. The brain was small and weighed 1080 gms. The calvarium was very thick and unusually heavy. It weighed 480 gms. Spinal cord showed no gross abnormalities. The heart was flabby, and weighed 250 gms.; mitral ring was very wide; the cusps were somewhat wrinkled and their edges were covered with tightly adherent vegetations; the aorta appeared normal. The left lung had a few apical adhesions, fibrotic tuberculosis without cavity formations. The thymus was not present. The thyroid was well developed. The left ovary showed cystic degeneration—a large reddish cyst with injected vessels; the left ovary itself was small and hypoplastic. The spleen was large and heavy. The liver and kidneys appeared grossly normal.

Microscopic examination showed much hyalinization atrophy and numerous fat cells in the muscular tissue. Bands of connective tissue, some delicate and some firm run throughout the muscular tissue. Thyroid gland had numerous follicles distended with smooth pink colored substance.

DISCUSSION

All four siblings were admitted to Wrentham State School because of mental retardation, lack of progress at school and poor social adjustment. Obvious physical handicaps were not present. Yet, although no muscular disability existed in childhood and the onset of myotonia dystrophica was recorded in Pearl at the age of 27, in Vincent at 17, in Tony at 35 and in Mary at 19, certain physical and neurological findings seem to indicate that mild subclinical changes existed long before myotonia dystrophica became clinically obvious. Sluggishness of knee-jerks was noticed on admission in Pearl, aged 17, Tony aged 12, and severe decrease of knee-jerks in Mary aged 10. Matron's complaints of laziness and unwillingness to work may well have been due to muscular difficulties which patients are unwilling to face or admit. The disease is of such slow insidious development that neither the patient nor the family are usually able to state definitely the time of the onset of the disease until myotonia or muscular dystrophy interferes with the patient's motion or capacity for work. All four siblings had physical stigmata recorded on admission, such as, high narrow arched palate, poorly formed ears, facial asymmetry, etc. None of the siblings had all the features characteristic for this disease, but as a group

Case #1. At age of 32 years

Case #2. At age of 29 years.

Case #3. At age of 28 years.

Case #4. At age of 17 years.

they had every symptom described in this condition. Two siblings had punctate opacities of the lens discovered with slit lamps; two had ptosis, one had diabetes, and one had excessive salivation; all had decreased deep tendon reflexes and normal superficial reflexes. Both boys had testicular atrophy and pronounced obesity. All siblings had high arched palate, high arched feet and severe hyperostosis frontalis and general thickening of calvarium. All had atrophy of hand muscles and all became slow, indolent, sleepy appearing and clumsy. Both boys had alopecia, especially pronounced in Vincent. All had monotonous nasal voices. All were mentally deficient. Mental defect in patients or siblings in families affected by myotonia dystrophica is common. According to Maas and Peterson[9], "mental changes almost always accompany much muscular wasting", and Billings and Ravin[10] found that "intellectually most of the cases were poorly endowed". Myotonia dystrophica is a familiar hereditary disease inherited as a dominant feature. Although it is said to affect males more than females, our patients were equally divided—two males and two females. The dystrophy of other organs besides muscles was very pronounced. It has been hypothesized by many writers that the disease is of endocrine origin.[11] The fronto-temporal baldness, the testicular atrophy, obesity, decreased 17-ketosteroids, hyperostosis interna of the frontal bones, structural cellular changes in pituitary and adrenals, pre-senile cataracts, are cited in favor of this theory. Of the siblings who died, Mary was 30, Vincent 43 and Tony 40 years old. The autopsy findings were: "fish-flesh" muscular changes typical for muscular dystrophy in Tony, changes in pituitary gland, hypoplasia and lipid paucity of adrenal cortex in all three patients; diffuse colloidal struma of the thyroid gland in both boys; well developed thyroid, cystic degeneration and hypoplastia of ovaries in Mary; hyperplastic thymus in Vincent and absent thymus in Mary and Tony.

These cases were differentiated from Thomsen's disease by the absence of myotonia in early childhood and the presence of muscular atrophy. They were differentiated from other muscular dystrophies by the distribution of the muscular atrophy involving mostly the small muscles of hands and feet, thighs and lower legs, myotonia, cataracts, characteristic baldness, gonadal atrophy, premature senility, and absence of contractures.

Microscopic picture of the muscles was typical for myotonia

dystrophica as described by Addie and Greenfield[1], Adams[5], Keschner[6], Black and Ravin[2], Hurvitz[7], Slauck[3], Wohlfart[4], Curschmann[8] and others. All stages from the early ones represented by densely packed long chains of central nuclei through hyalinization and homonization of fibers, especially pronounced in Mary, the splitting of fibers, Heidenhain's Ringebinde, to completely atrophic fibers represented by clumps of darkly stained nuclei, and separated by fat and connective tissue were seen. Smooth muscles were not affected. No mitotic figures were found. Tubular atrophy of testicles, cellular changes in pituitary with increase in basophilic cells, congenital vascular malformation of hemangioma type in the white matter of the right frontal lobe, hyperactive thyroid in two patients, cataracts showed the extent of extra-muscular dystrophy.

SUMMARY

Four cases of myotonia dystrophica in two brothers and two sisters with description of three autopsies are presented. The impression that the cases of myotonia dystrophica are of endocrine origin is supported by these cases.

REFERENCES

1. Addie, W. J. and Greenfield, J. G.: Brain, 46:73-127, 1923.
2. Black, W. C. and Ravin, A.: Studies in Dystrophia Myotonica, Arch. Neurol. & Psychiat., 44:176-191, 1947.
3. Slauck, A.: Pathologische Anatomie de Myopathien. In Bumke und Foerster: Handbuch der Neurologie, 16:412-431, 1936.
4. Wohlfart, G.: Dystrophia Myotonica and Myotonia Congenita. Histopathological Studies with Special Reference to Changes in Muscles. J. Neuropath & Exper. Neurol., 10:109-124, 1951.
5. Adams, R. D.; Denny-Brown, D. and Person, C. M.: Diseases of the Muscle. A Study in Pathology.
6. Keschner, M. and Davison, C.: Dystrophia Myotonica. A Clinico-pathologic Study. Arch. Neurol. & Psychiat., 30:1259-1275, 1933.
7. Hurwitz, S.: Primary Myopathies. Report of 36 Cases and Review of the Literature. Arch Neurol. & Psychiat., 36:1294-1316, 1936.
8. Curschmann, H.: Zur Nosologie und symptomatologie der myotonischen Dystrophie. Deutsche Arch. f. klin. Med., 149:129, 1925.
9. Maas, O. and Peterson, A. S.: Mental Changes in Families Affected with Dystrophia Myotonica. Lancet, 1:24, 1937.
10. Billings, E. G. and Ravin, A.: A Psychiatric Study of Patients Manifesting Dystrophia Myotonica. Am. J. of Psychiat., 97:1116, 1941.
11. Janney, N. W.; Goodhart, S. P. and Isaacson, V. I.: The Endocrine Origin of Muscular Dystrophy. Arch. Int. Med., 21:188.
12. Heidenhain, M.: Ueber progressive Veranderungen der Muskulatur bei Myotonia atrophica. Beitr. z. path. Anat. u.z. allg. Path., 64:198, 1918.

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DEPARTMENT OF ABSTRACTS

Conducted by
MICHAEL A. BRESCIA, M.D., NEW YORK

SHAFFER, T. E.; SYLVESTER, R. F.; BALDWIN, J. N. and RHEINS, M. S.: STAPHYLOCOCCAL INFECTIONS IN NEWBORN INFANTS. II. REPORT OF 19 EPIDEMICS CAUSED BY AN IDENTICAL STRAIN OF STAPHYLOCOCCUS PYOGENES. (American Journal of Public Health, 47:990, August 1957).

Since 1954 there has been an increasing incidence throughout the world of epidemic infections in hospital nurseries for the newborn, the etiological agent in the great majority of these epidemics being penicillin-resistant S. pyogenes, phage type 42B/47C/44A/52/80/81. Bacteriophage typing and determination of antibiotic sensitivity are shown to be valuable technics for the epidemiologic study of staphylococcal infections. Control of outbreaks in nurseries depends on prevention of nasal colonization of infants. Prophylactic medication with an appropriate antibiotic offers a practical method for preventing nasal colonization. Nasal carriage precedes clinical infection in these epidemics. The majority of infants who develop infection do so after discharge from the hospital. Staphylococcal disease in infants under two months of age and mastitis in nursing mothers should be reportable diseases so that public health authorities can investigate hospital nurseries as possible sources of infection. *Authors' Summary.*

PEW, W. L.: NEUROLOGIC MANIFESTATIONS OF INFECTIOUS MONONUCLEOSIS. (Northwest Medicine, 56:695, June 1957).

Infectious mononucleosis is a systemic disease which affects the nervous system in a significant number of cases. Any form or degree of nervous system involvement may be seen. This disease should be suspected in any unexplained neurologic disease. Complete, rapid recovery is the rule but severe, permanent residuals or death may result. In thirty-two consecutive cases of I. M. in children 16 or younger, six had definite neurologic complications and six had suggestive history, signs or symptoms. One patient died and another was left with irreversible brain damage.

Author's Summary.

LANGE, K.: THE IMMUNOLOGIC BASIS OF GLOMERULONEPHRITIS AND THE NEPHROTIC SYNDROME. (New York State Journal of Medicine, 57:2095, June 15, 1957).

Acute glomerulonephritis and the nephrotic syndrome are probably the result of a complement-binding antigen-antibody reaction. The glomerular membrane is altered by coating with an antigen to which antibodies are formed by the patient. When these antibodies appear in the circulation, they join with the sessile antigen in the glomeruli, and a tissue-destroying, complement-binding, antigen-antibody reaction results. Based on these immunologic considerations and on the fact that corticosteroids depress antibody formation, a therapeutic regime for the treatment of the nephrotic syndrome was developed. This therapeutic regime has led to a reduction in mortality in this disease in a ratio of 12.8:1 during a six year observation period. Future therapeutic approaches to glomerulonephritis and nephrosis should consider the immunologic background of these diseases.

AUTHOR'S SUMMARY.

HORNER, F. A.; STREAMER, C. W.; CLADER, D. E.; HASSELL, L. L.; BINKLEY, E. L., JR. AND DUMARS, K. W., JR.: EFFECT OF PHENYLALANINE-RESTRICTED DIET IN PHENYLKETONURIA II. (A.M.A. Journal of Diseases of Children, 93:615, June 1957).

Six patients with phenylketonuria have been under treatment with phenylalanine-restricted diets for periods varying from 8 to 22 months. Two children who were not treated until the fifth year of life are showing sustained increments in their intellectual functioning. Of the two children treated in the third year of life, one is showing marked acceleration in her developmental pattern while the second shows little significant change after 14 months of treatment. Two newborn infants treated since the appearance of phenylketonuria are following a healthy developmental pattern after 21 and 7 months respectively. It is suggested that on the basis of these observations the treatment of older phenylketonurics may prove valuable in some cases. The treatment of newborns with phenylketonuria may prevent the characteristic and profound mental changes often associated with this abnormality. It is possible that negative urinary tests for phenylketonuria may be misleading during the neonatal period.

AUTHORS' SUMMARY.

LITCHFIELD, H. R.: CLINICAL EVALUATION OF MEPROBAMATE IN DISTURBED AND PREPSYCHOTIC CHILDREN. (Annals of New York Academy of Sciences, 67:828, May 9, 1957).

Meprobamate is the drug of choice in the treatment of a wide variety of children of all ages manifesting prominent symptoms of anxiety and tension, with or without organic disease, who are seen and treated primarily by the practicing pediatrician on an ambulatory outpatient basis. The drug appears to be of definite value in the control of petit mal epilepsy. It produces a tranquilizing effect without dulling of consciousness, disturbance of the mental faculties, or hypnosis. In no instance did this medication, in young or older children, retard their mental abilities. It appears to be essentially nontoxic. M.A.B.

DUBROW, E.: ENCHANTMENT OF CONVENTIONAL THERAPY FOR APPETITE STIMULATION. NEW TREATMENT. (Quarterly Review of Pediatrics, 12:142, August 1957).

Three groups of underweight children with poor appetites were given a mixture of reserpine (.05 mg.), vitamin B1 (10 mg.) and vitamin B12 (10 mg., crystalline). All three groups exhibited statistically significant weight gains and demonstrated improved appetite. The most striking gains were in those children who were tense, nervous and high-strung. Many of these children were calmer and better adjusted emotionally after treatment. No side effects were noted with the dosage used. *Author's Summary.*

HAND, J. R.: TREATMENT OF UNDESCENDED TESTIS AND ITS COMPLICATIONS. (Journal American Medical Association, 164: 1185, July 13, 1957).

Changes caused by the failure of a testis to descend per se do not impair fertility if the testis reaches the scrotum by puberty. In a small series of cases, bilateral spontaneous descent gave a slightly higher percentage of fertility than bilateral orchipexy at the same age. This speaks in favor of giving the testes a chance to descend spontaneously. Clinical signs of possible spontaneous descent are (1) mobility in the normal line of descent, (2) palpation of a small cord, (3) responses of the scrotum when the cremaster reflex is initiated, (4) a patent scrotel neck, and (5) absence of a demonstrable hernia. Adequate hormone therapy will hasten descent, provided there is no mechanical obstruction or agenesis. Such

therapy is of particular value in boys with bilateral retention. It should not be used when there is a concurrent hernia or torsion, when the testis is arrested in an ectopic position, or, when the patient has passed puberty. It is best to defer hormone stimulation until the age of 8 years. This allows (1) more time for possible spontaneous descent but still enough for orchipexy prior to puberty if descent has not been induced, and (2) the development of a larger testis, which permits better fixation than can be accomplished in the younger child and thus lessens the possibility of postoperative retraction. Neither hormone therapy nor operative intervention should be deferred beyond puberty. In the evaluation of fertility, consideration must be given not only to scrotal residence by puberty but to the quality of the testis. This can be accurately determined only by biopsy. *Author's Summary.*

TALBOT, N. B.; CRAWFORD, J. D. and COOK, C. D.: APPLICATION OF HOMEOSTATIC PRINCIPLES TO THE MANAGEMENT OF NEPHROTIC PATIENTS. (New England Journal of Medicine, 256: 1080, June 6, 1957).

Metabolic studies are presented indicating that edema children with the nephrotic syndrome may be the product of a series of reactions to the primary pathological loss of plasma albumin into the urine. In order of occurrence, these reactions are thought to be hypoalbuminemia, vascular hypovolemia, conservation of sodium and companion anions by the kidneys under the controlling influence of the body's sodium homeostatic system, increase in body solute (sodium and so forth) content, conservation of a physiologically equivalent volume of water by the kidneys under the controlling influence of the body's "water-concentration" homeostatic system, and finally, accumulation of edema fluid in extracellular spaces. The value of a variety of therapeutic procedures for the control of such edema is considered in the light of this thesis. It is concluded that corticosteroid therapy occupies a unique position in this list because it acts to suppress the lesion primarily responsible for the development of the edema. Because of this action, it should probably be given beyond the time when edema disappears and until the nephrotic process has become either totally quiescent or maximally ameliorated by corticosteroids.

Authors' Summary.

JOHNSON, A. M. and ROBINSON, D. B.: THE SEXUAL DEVIANT (SEXUAL PSYCHOPATH)—CAUSES, TREATMENT, AND PREVENTION. (Journal American Medical Association, 164:1559, August 3. 1957).

Clinical research reveals that destructive, direct sexual acting-out and perverse sexual behavior are forms of sexual deviation that develop in most instances from unconscious, or, less frequently, conscious pathological fostering of deviant sexual behavior early in life within the family setting. Investigation further demonstrates that, engrafted upon this framework, direct and overt sexual seductive behavior inclines the child toward later direct, sexual, destructive acting-out, while indirect, disguised sexual stimulations engender the sexual perversions. Since therapy of the full-blown sexual deviant is long and difficult, prophylaxis is the preferred attack on the problem. A first approach consists of education of the parents. Many well-intentioned parents, influenced by the misinterpretations of Freud's theories, do not observe proper and healthy rules of modesty in the home. Dangerously little knowledge has led them to believe that "neurosis is based on repression of sexual curiosity and interests", and, therefore, it is erroneously believed that an open approach to sexuality provides an emotionally healthful atmosphere. The resulting multiple forms of bodily exposure may result in unwitting and excessive overstimulation of the child. Second, pediatricians and family physicians must squarely face the unequivocal truth that all degrees of seduction and sexual stimulation occurs all too commonly, and in families which exhibit every outward aspect of respectability, decency, and conformity with convention. Equipped with understanding and employing relatively simple procedures, the family physician and the pediatrician may well reduce significantly the incidence of sexual deviations.

M.A.B.

WOOD, N. and O'GORMAN, G.: ERYTHROMYCIN TREATMENT OF A DIPHTHERIA CARRIER EPIDEMIC IN A MENTAL DEFICIENCY HOSPITAL. (Antibiotic Medicine and Clinical Therapy, 4:465, August 1957).

During a period of eight weeks, following the isolation of virulent Corynebacterium diphtheriae type mitis from the throat of a child who had died from acute respiratory obstruction, two patients with borderline cases and twenty-nine carriers of virulent C.

diphtheriae type mitis were recognized by repeated mass nose and throat swabbings in a crowded mental hospital housing 164 patients. This carrier epidemic was controlled by segregation within the hospital and by the oral administration of erythromycin to all patients with positive cultures. During the following seven months, only one of those treated again became positive, and it is suggested that this was a reinfection rather than a relapse. In similar closed communities, routine swabbing of sore throats and nasal discharges may reveal an unsuspected concentration of diphtheria infection. *Authors' Summary.*

LARKIN, V. D.: POLYOXETHYLENE DODECANOL VAPORIZATION IN THE TREATMENT OF RESPIRATORY INFECTIONS OF INFANTS AND CHILDREN. (New York State Journal of Medicine, 57:2667, August 15, 1957).

Tablets containing polyoxyethylene docecanol (PED) and aromatics are nontoxic and nonirritating when one tablet is used per 8 ounces of water in an electric steam vaporizer for a continuous treatment of eight hours exposure to the vapors. Patients subjected to twice and five times this dosage did not display any indication of toxicity or irritation. Steam treatments with tablets containing PED had a rapid, striking effect in eliminating or substantially reducing cough regardless of the severity of the underlying respiratory infection. No other forms of therapy, including cough syrups, antibiotics, or vaporizers, medicated or not, approached the effectiveness of the PED tablets in this respect. Respiratory distress, including dyspnoea and wheezing showed marked improvement with PED steam therapy much more rapidly and completely than with any other therapy used.

Although all cases of croup responded favorably with PED steam treatment, the number of cases is too small to justify a valid comparison with steam treatment alone. Patients with infirmation of the tonsils or pharynx, or, with nasal congestion or discharge did not obtain any significant therapeutic effect from treatment with PED tablets in a steam vaporizer. *Author's Summary.*

BOOK REVIEWS

Conducted by
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J.A.M.A. CLINICAL ABSTRACTS OF DIAGNOSIS AND TREATMENT. Edited by I. Phillips Frohman, M.D. Cloth. Pp. 564. Price $5.50. New York and London: International Medical Book Corporation with Grune & Stratton, Inc., 1957.

This is another in a series of books carrying some of the abstracts that previously appeared in the "Medical Literature Abstracts" section of the J.A.M.A. These are always helpful books and under one cover there is a large number of articles of importance from the world literature which seems to be growing greater and greater. I still feel as I noted in reviewing the first such volume that a cumulative index would greatly enhance the value of these books of abstracts. The index itself might become too cumbersome, in which case a separate index volume would be in order, approximately every five years. This book differs from the previous volumes in that the abstracts are segregated as to systems rather than as to specialties. This pattern seems better and should have a wider appeal. There are some seventy abstracts under the different systems that are of pediatric interest and these can be found easily by referring to the index. These books have merit and deserve continuance of publication. M.A.B.

CLINICAL PSYCHOLOGY OF EXCEPTIONAL CHILDREN. By C. M. Louttit, M.D., et al. Cloth. Pp. 573. Price $6.00. New York: Harper & Brothers, 1957.

In the two previous editions of this widely used text book, published in 1936 and in 1947, emphasis was placed on deviant behavior of children. The third edition also has as its central concern the behavior problems of children who deviate from the norm, but, in the author's words, "The discussion of such problems would be from the point of view of the clinical psychologist." Special attention is directed to several groups of exceptional children and a separate chapter is devoted to each; included are those related to ability, both retarded and gifted, and those who have difficulty with school progress or with special school subjects. Two excellent

chapters follow dealing with children who show deviation in social behavior and personality. The final chapters are devoted to speech, sensory and neurological defects and physical disabilities, and their relationship to psychological problems. The foregoing represent major changes in the third edition. Chapters in earlier editions dealing with personality and conduct problems have been replaced with sections which provide background, conditions, influences, detection, treatment and correction of the problem behavior under consideration. Treatment of the special conditions is dealt with separately. An additional chapter summarizes normal development, replacing an earlier one on the nature of problem behavior. The third edition of this text can now be used as a handbook or manual, since each chapter summarizes the area concerned and is replete with charts, case-history material, vast bibliographies and other illustrative material. Dr. Louttit invited five younger colleagues to contribute chapters on topics concerned with their special interest and training, and among these are Michael M. Reece, Victor and Mildred G. Goertzel. In the opinion of this reviewer, a most worthwhile chapter is the one dealing with neurological and physical disabilities and the important part they play in the etiology of psychological problems. Here, the contributor, Dr. David H. Crowell of the University of Hawaii, has differentiated conditions based on neural pathology, orthopedic or neuromuscular disabilities and systemic conditions. ELEANOR ROTHENBERG, M.A.

THE COMPLEAT PEDIATRICIAN. 7th Ed. By Wilburt C. Davison, M.A., D.Sc., LL.D., M.D. and Jeane Davison Levinthal, B.A., M.D. Paper. Pp. 300 plus. Price $4.50. Durham, N. C.: Seeman Printery for Duke University Press, 1957.

On the title page the book is said to be for the use of general practitioners, pediatricians, interns and medical students and so it can very well be. This type of book should have a wide appeal because it gives a quick and brief answer to practically the entire pediatric field. It is ideal for an easy reference source but for its fine print would have greater appeal to the intern and student for quick "boning up". It is amazing how much valuable material can be contained in such a comparatively small volume. The many tables enhance the value of the book. It is the type of book one keeps at hand, on the desk, for quick and easy reference. M.A.B.

THE METABOLIC RESPONSE TO NEONATAL SURGERY. By P. P. Rickham, M.S., F.R.C.S., D.C.H. Cloth. Pp. 93. Illustrated. Price $5.00. Cambridge, Mass.: Harvard University Press for the Commonwealth Fund, 1957.

This is a book that all pediatricians will want to read and digest and certainly would be of great benefit to the surgeon who may be called upon to operate on the newborn. The author has recorded in detail the electrolyte and other pertinent findings in a newborn subjected to minor and major surgical procedures. This detailed study included nine subjects but the discussion and recommendations were not only based on the experience with these nine cases but some 300 other surgical procedures on newborns who, however, were not subjected to such a detailed metabolic study. In his conclusion the author states: "how little of our pre- and post-operative treatment is based on scientific facts and how much of it is purely empirical. We know as yet very little about the many physiological and pathological changes which we initiate when operating on newborn infants. There is no doubt that without greater insight into the physiology of the newborn infant, and the processes by which he adjusts himself from parasitic intra-uterine existence to separate external life, there will be little further progress in neonatal surgery". The author, nevertheless, has made an excellent contribution to the understanding of the newborn. It behooves the surgeon and the pediatrician who might be partially responsible for the post-operative care of a newborn to be as cognizant as possible of the metabolic processes of the newborn to save an otherwise fine surgical procedure from going to naught. M.A.B.

Complications of Hiatal Hernia. A. Mazzarella and C. Pototschnig. (Minerva pediat., 8:84-89, Feb. 4, 1956). The authors report on a case of hiatal hernia with esophageal stricture in a 15-month-old infant. The condition was first believed to be a stenosis of the esophagus. From the age of 3 months the infant showed some regurgitation during meals. After the age of 8 months regurgitation and vomiting occurred in cycles. At operation a diaphragmatic hiatal hernia and stenosis of the cardia located 3 cm. above the diaphragm were found. Forty-eight hours after the operation the child was given food and no regurgitation or vomiting occurred.—J.A.M.A.

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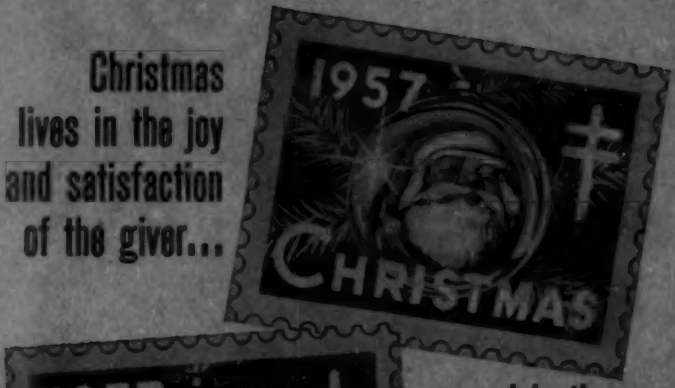

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